

Amendment under Art.34

31. (Add) An agent for inhibiting the phosphorylation of c-Jun, wherein the agent comprises one or more peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase 3 as the active ingredients:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 2 in Sequence List,

(v) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 3 in Sequence List, wherein the peptide is human originated,

(vi) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide comprising the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(x) a peptide having mutations of one to several amino acid residues in

an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xi) a peptide having mutations of one to several amino acid residues in an amino acid sequence of the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xiii) a peptide having a homology of 70% or more to peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xiv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 4 in Sequence List, or

(xv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 5 in Sequence List.

32. (Add) An agent for inhibiting the ability to activate transcription of c-Jun, wherein the agent comprises one or more peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase 3 as the active ingredients:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 2 in Sequence List,

(v) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 3 in Sequence List and human originated,

(vi) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide comprising the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(x) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xi) a peptide having mutations of one to several amino acid residues in an amino acid sequence of the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xiii) a peptide having a homology of 70% or more to peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the

phosphorylation of c-Jun by JNK3,

(xiv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 4 in Sequence List, or

(xv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 5 in Sequence List.

33. (Add) a pharmaceutical composition comprising an effective dose of an agent for inhibiting the ability to activate transcription of c-Jun according to Claim 32.

34. (Add) The pharmaceutical composition according to Claim 33, wherein the pharmaceutical composition is an agent for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun by c-Jun N-terminal kinase 3.

35. (Add) A pharmaceutical composition comprising an effective dose of one or more peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase 3 (JNK3), wherein the pharmaceutical composition is an agent for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun by JNK3:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 2 in Sequence List,

(v) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 3 in Sequence List and human originated,

(vi) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide comprising the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(x) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xi) a peptide having mutations of one to several amino acid residues in an amino acid sequence of the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xiii) a peptide having a homology of 70% or more to peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xiv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 4 in Sequence List, or

(xv) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 5 in Sequence List.

36. (Add) A pharmaceutical composition comprising an effective dose of one or more polynucleotides encoding at least one peptide selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase (JNK3), wherein the pharmaceutical composition is an agent for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun caused by JNK3:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 2 in Sequence List,

(v) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 3 in Sequence List and human originated,

(vi) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide comprising the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by

JNK3,

(x) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xi) a peptide having mutations of one to several amino acid residues in an amino acid sequence of the peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xiii) a peptide having a homology of 70% or more to peptide of said (v), wherein the peptide is human originated and has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(xiv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 4 in Sequence List, or

(xv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 5 in Sequence List.

37. (Add) The pharmaceutical composition according to at least one of Claims 34 to 36, wherein the disease caused by the phosphorylation of c-Jun by c-Jun N-terminal kinase 3 is a neurodegenerative disease.

38. (Add) The pharmaceutical composition according to Claim 37, wherein the neurodegenerative disease is polyglutamine disease, Huntington's disease, spinocerebellar ataxia, bulbospinal muscular atrophy, dentatorubropallidoluysian atrophy, Alzheimer's disease, Down's disease, Parkinson's disease, Lewy body dementia, multiple system atrophy, familial amyotrophic lateral sclerosis,

progressive supranuclear palsy, corticobasal degeneration, Pick's disease, familial British dementia, Creutzfeldt-Jakob disease, Gerstmann-Sträussler syndrome, mad cow disease (bovine spongiform encephalopathy) (BSE), or familial dementia accompanying neuroserpin inclusion bodies.

39. (Add) An agent for inhibiting phosphorylation of c-Jun, wherein the agent comprises one or more peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase 3 as the active ingredients:

- (i) BMAL1,
- (ii) BPL1,
- (iii) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 1 in Sequence List,
- (iv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 2 in Sequence List,
- (v) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,
- (vi) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,
- (vii) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,
- (viii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,
- (ix) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 4 in Sequence List, or

(x) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 5 in Sequence List.

40. (Add) A method for inhibiting the phosphorylation of c-Jun, wherein the method comprises putting one or more peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase 3 (JNK3) in coexistence with JNK3:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 2 in Sequence List,

(v) a peptide comprising at least one of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vi) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 4 in Sequence List, or

(x) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 5 in Sequence List.

41. (Add) The method for inhibiting the phosphorylation of c-Jun according to Claim 40, wherein the peptide group consists of (i), (ii), (iii), (iv), (ix), or (x).

42. (Add) An agent for inhibiting the ability of c-Jun to activate transcription, wherein the agent comprises one or more than two peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase 3 as the active ingredients:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 2 in Sequence List,

(v) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vi) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 4 in Sequence List, or

(x) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 5 in Sequence List.

43. (Add) A method for inhibiting the ability of c-Jun to activate transcription , wherein the method comprises putting one or more peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase 3 (JNK) in coexistence with JNK3:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 2 in Sequence List,

(v) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vi) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 4 in Sequence List, or

(x) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 5 in Sequence List.

44. (Add) A pharmaceutical composition comprising an effective dose of an agent for inhibiting phosphorylation of c-Jun according to Claim 39, or an agent for inhibiting the ability of c-Jun to activate transcription according to Claim 42.

45. (Add) A pharmaceutical composition according to Claim 44, wherein the pharmaceutical composition is an agent for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun by JNK3.

46. (Add) A pharmaceutical composition comprising an effective dose of one or more peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase (JNK3), wherein the pharmaceutical composition is an agent for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun by JNK3:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 2 in Sequence List,

(v) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vi) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide having mutations of one to several amino acid residues in

an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 4 in Sequence List, or

(x) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 5 in Sequence List.

47. (Add) A pharmaceutical composition comprising an effective dose of one or more polynucleotides encoding at least one peptide selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase (JNK3), wherein the pharmaceutical composition is an agent for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun by JNK3:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 2 in Sequence List,

(v) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vi) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 4 in Sequence List, or

(x) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 5 in Sequence List.

48. (Add) The pharmaceutical composition according to at least one of Claims 45 to 47, wherein the disease caused by the phosphorylation of c-Jun by c-Jun N-terminal kinase is a neurodegenerative disease.

49. (Add) The pharmaceutical composition according to Claim 48, wherein the neurodegenerative disease is polyglutamine disease, Huntington's disease, spinocerebellar ataxia, bulbospinal muscular atrophy, dentatorubropallidoluysian atrophy, Alzheimer's disease, Down's disease, Parkinson's disease, Lewy body dementia, multiple system atrophy, familial amyotrophic lateral sclerosis, progressive supranuclear palsy, corticobasal degeneration, Pick's disease, familial British dementia, Creutzfeldt-Jakob disease, Gerstmann-Strässler syndrome, mad cow disease (bovine spongiform encephalopathy) (BSE), or familial dementia accompanying neuroserpin inclusion bodies.

50. (Add) A method for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun by JNK3, wherein the method comprises putting one or more peptides selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase (JNK3) in coexistence with JNK3:

(i) BMAL1,

(ii) BPL1,

(iii) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 1 in Sequence List,

(iv) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 2 in Sequence List,

(v) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vi) a peptide comprised of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(vii) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(viii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,

(ix) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 4 in Sequence List, or

(x) a peptide comprised of the amino acid sequence represented by SEQ

ID NO: 5 in Sequence List.

51. (Add) A method for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun by JNK3, wherein the method comprises utilizing one or more polynucleotides encoding at least one peptide selected from the following peptide group having a function for interacting with c-Jun N-terminal kinase (JNK3) to express the peptide encoded by the polynucleotides, wherein the

peptide inhibits the phosphorylation of c-Jun by JNK3:

- (i) BMAL1,
- (ii) BPL1,
- (iii) a peptide composed of the amino acid sequence represented by SEQ ID NO: 1 in Sequence List,
- (iv) a peptide composed of the amino acid sequence represented by SEQ ID NO: 2 in Sequence List,
- (v) a peptide comprising at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,
- (vi) a peptide composed of at least 5 consecutive amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,
- (vii) a peptide having mutations of one to several amino acid residues in an amino acid sequence of at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,
- (viii) a peptide having a homology of 70% or more to at least one peptide of said (i) to (iv), wherein the peptide has a function for inhibiting the phosphorylation of c-Jun by JNK3,
- (ix) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 4 in Sequence List, or
- (x) a peptide comprised of the amino acid sequence represented by SEQ ID NO: 5 in Sequence List.

52. (Add) A method for preventing and/or treating disease(s) caused by the phosphorylation of c-Jun by JNK3, wherein the method comprises using a pharmaceutical composition according to at least one of Claims 44 to 49.

53. (Add) The method for preventing and/or treating disease(s) according to at least one of Claims 50 to 52, wherein the disease caused by the phosphorylation of c-Jun by c-Jun N-terminal kinase 3 is a neurodegenerative disease.

54. (Add) The method for preventing and/or treating disease(s) according to Claim 53, wherein the neurodegenerative disease is polyglutamine disease, Huntington's disease, spinocerebellar ataxia, bulbospinal muscular atrophy, dentatorubropallidoluysian atrophy, Alzheimer's disease, Down's disease, Parkinson's disease, Lewy body dementia, multiple system atrophy, familial amyotrophic lateral sclerosis, progressive supranuclear palsy, corticobasal degeneration, Pick's disease, familial British dementia, Creutzfeldt-Jakob disease, Gerstmann-Strässler syndrome, mad cow disease (bovine spongiform encephalopathy) (BSE), or familial dementia accompanying neuroserpin inclusion bodies.